Congenital absence of the portal vein (CAPV) is a rare anomaly where the intestinal and splenic venous drainage enters the various systemic veins through a congenital portosystemic shunt. Recently, this anomaly has been increasingly recognized on non-invasive imaging studies, especially in pediatrics [1-3]. In this report, we describe abdominal computed tomography (CT) and brain magnetic resonance image (MRI) findings of this rare anomaly in a middle-aged man that presented with portosystemic encephalopathy.

Case Report

A 39-year-old man presented to our emergency room with consciousness disturbance and behavior change. The patient had no relevant family, medical or surgical history. A neurological examination showed disorientation and impairment of intellectual function. Laboratory results revealed mild elevation of the level of AST (38 U/L), total bilirubin (1.3 mg/dL) and marked elevation of ammonia (138 μmol/L). All viral markers were negative and the other parameters were within normal limits. Brain MRI showed bilateral high signal intensities in the globus pallidus representing characteristic hepatocerebral degeneration (Fig. 1A). A subsequent abdominal CT image demonstrated the absence of the portal vein and portosystemic shunt between the inferior mesenteric vein (IMV) and left internal iliac vein (Fig. 1B-F). Varicose veins in the pelvic cavity were also seen (Fig. 1F). A markedly distended IMV joined the superior mesenteric vein (SMV) just below the confluence of the SMV and splenic vein. Two small hepatic cysts were seen in the left lobe, but no solid mass was identified in the liver. The hepatic arteries, inferior vena cava (IVC) and left internal iliac vein were diffusely distended. Signs of portal hypertension such as collateral veins, ascites and splenomegaly were not seen. No other anomalies were present in the patient. After conservative management for two days, the level of serum ammonia became normalized, as did consciousness and the behavior of the patient. Currently the patient is undergoing routine follow-up.

Discussion

The portal vein develops between the fourth and tenth
Fig. 1. A 39-year-old man with congenital absence of the portal vein with a portosystemic shunt into the left internal iliac vein through the dilated IMV.

A. T1 weighted MRI shows bilateral high signal intensity in both globus pallidus (arrows) representing typical hepatocerebral degeneration.

B. An abdominal CT scan shows absence of the portal vein. The splenic vein (arrowheads) and distended hepatic arteries (arrows) are seen.

C. The IMV (arrows) joins the SMV (asterisk) just below the confluence of the splenic vein and SMV.

D, E. The distended IMV (small arrows) descends into the pelvic cavity and drains into the left internal iliac vein (large arrow).

F. The shaded surface display shows the confluence between the splenic vein (arrowheads) and SMV (asterisk) which drains into the left internal iliac vein (large arrow) through the dilated IMV (small arrows).
weeks. It forms by involution of the paired vitelline veins. Excessive involution can result in congenital absence of the portal vein [4, 5] and as a result, a congenital portosystemic shunt into the various systemic veins occurs.

Subsequent to the first report by Abernethy in 1793 [1, 5], the presence of CAPV has been increasingly reported, especially in pediatric patients [1, 2]. Most of the cases of CAPV occur in females and children, but cases in males and adults have been documented recently [4, 5]. CAPV has been classified into two types according to the absence (type a) or presence (type b) of the confluent superior mesenteric and splenic veins (6). The present case belongs to type b with confluence of the SMV and splenic vein.

The most common drainage site of the portal vein is the IVC (5, 7), but drainage into the renal vein, left hepatic vein, right arium and azygos vein has been reported [7]. Our case shows an extrahepatic portosystemic shunt that drains into the left internal iliac vein through the IMV.

Hepatic lesions associated with CAPV have been well recognized. Focal nodular hyperplasia is the most common hepatic lesion associated with CAPV and case reports of a hepatoblastoma, hepatic adenoma, hepatocellular carcinoma and nodular regenerative hyperplasia associated with CAPV have been documented [7]. Congenital heart disease such as atrial septal defect, patent ductus arteriosus and ventricular septal defect have been frequently reported with CAPV [7, 8]. Polysplenia, Goldenhar syndrome, skeletal malformations and multicystic dysplastic kidney have been also described [7]. Our case showed no solid liver lesions or other congenital anomalies.

Because of the extrahepatic portosystemic shunt, neurological symptoms representing portosystemic encephalopathy (PSE) are expected to occur, but CAPV with PSE has been reported only in few cases [8-10]. It is generally believed that chronic exposure to hyperammonemia may increase the possibility of PSE. Most cases of reported CAPV have been in children, and it is believed that these patients are not old enough to have portosystemic encephalopathy [7, 9]. However, Wakamoto et al. reported subclinical PSE presenting with white matter atrophy and ventricular enlargement in a child with CAPV [9]. Thus, in patients with CAPV, whether symptoms are present or not, a careful neurological evaluation is needed to improve the prognosis [4, 9]. Our patient showed hyperammonemia, consciousness disturbance and behavioral change. MRI demonstrated bilateral high signal intensity in both globus pallidus on a T1-weighted image representing typical hepatocerebral degeneration. Long-term exposure to hyperammonemia in this patient might have induced the underlying condition. Liver transplantation should be considered for the treatment of symptomatic patients that do not respond to medical therapy [4, 10].

In conclusion, CAPV is a rare congenital anomaly that has an extrahepatic portosystemic shunt and can cause portosystemic encephalopathy.

References
Kum Rae Kim, et al: Congenital Absence of the Portal Vein in an Adult Man Presenting with Portosystemic Encephalopathy

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© 2008;58:155-158

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Since portal vein is a major blood vessel (congenital absence of the portal vein), it is closely related to the development of the liver and pancreatic ducts. Congenital absence of the portal vein is associated with portosystemic encephalopathy (portosystemic encephalopathy), which indicates the development of liver cirrhosis which is also associated with portal hypertension.

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